Congenital Heart Disease In Children

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THE PHYSICIAN RENDERING primary care has a significant role to play in the management of children with congenital heart disease. He decides when a child should be referred to a pediatric cardiologist or center for definitive diagnostic tests and reviews their recommendations with the parents in order to implement an optimal therapeutic regimen. This necessitates a background of basic physiology and the diagnostic features of the common congenital cardiac lesions, as well as current knowledge of the natural history and results of surgical intervention. His role should be active rather than passive.

The most common lesions seen in older children include ventricular and atrial septal defects (VSD, ASD), patent ductus arteriosus (PDA), pulmonic stenosis with intact septum, tetralogy of Fallot, coarctation of the aorta and aortic stenosis. More complicated lesions are rare, and management has usually begun in infancy.

It is convenient to categorize patients into severity groups dependent upon the natural history and results of operation—for example, mild (excellent prognosis, operation not required), moderate (expert opinions differ, operation dependent upon evaluation of patient's general status), severe (operation definitiely indicated), inoperable (surgical mortality unacceptable).

Some lesions result in an increased volume of blood presented to the ventricle resulting in "volume work" (VSD, ASD, PDA) while in others, the ventricle must perform increased "pressure" work to overcome resistance (pulmonic or aortic stenosis, coarctation of aorta, left-to-right shunts complicated by pulmonary vascular obstructive disease).

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TABLE 1.—*Ventricular Septal Defects Severity Classification*

	Q_p/Q_s	P_p/P_s
Mild	<1.5	<0.5
Moderate	1.4 to 2.2	<0.5
Severe	>2.2	>0.5
Inoperable	<1.5	1.0

Q_p=Pulmonary Blood Flow. Q_s=Systemic Blood Flow. P_p=Pulmonary Artery Pressure. P_s=Systemic Blood Pressure.

Ventricular Septal Defect

Typical findings in VSD include a thrill in the fourth left interspace and a loud, rough, holosystolic murmur in that area. The natural history of this defect varies with the severity of the hemodynamic abnormality. The clinical course is dependent on the magnitude of pulmonary blood flow and reactivity of the pulmonary vascular bed. The severity of the defects are classified in Table 1.

Indications for early cardiac catheterization are exercise intolerance, cardiac enlargement, congestive heart failure, or evidence of right ventricular enlargement. If none of these problems is present, diagnostic studies are done electively after five years of age. Operation is curative unless the defect is incompletely closed, or heart block is induced.

Atrial Septal Defect

Classical findings in atrial septal defect are a parasternal impact, an ejection systolic murmur at the second left interspace, a widely split fixed S_2, and electrocardiogram evidence of right ventricular enlargement or rR′ pattern in the right precordial leads with roentgen evidence of cardiomegaly and increased pulmonary vascular markings. Operation is indicated unless the defect is very small and the ratio of pulmonic to systemic blood flow is less than 1.5, or severe pulmonary vascular obstruction has resulted in a right-to-left shunt with cyanosis. Surgical mortality in operable patients is less than one percent, and the result is curative.

Patent Ductus Arteriosus

Patent ductus arteriosus is characterized by a crescendo systolic-decrescendo diastolic machinery murmur. Cardiac catheterization is indicated only if the murmur is atypical, or there is evi-

dence of right ventricular enlargement (RVE). Operation is indicated in symptomatic children and, electively, in all patients at about four years of age. Operative mortality is less than one percent. Correction is contraindicated if there is cyanosis due to a right-to-left shunt.

Pulmonic Stenosis

The findings in pulmonic stenosis include a thrill and a rough ejection systolic murmur in the second left interspace. The lesion is characterized as mild if the peak systolic gradient from right ventricle to pulmonary artery is less than 50 mm of mercury, and severe if the peak systolic gradient is greater than 80 mm Hg. Gradients from 50 to 80 mm of mercury are classified as being moderate. Cardiac catheterization is performed electively after five years of age and earlier if there is clinically detectable cyanosis, cardiac enlargement or ECG evidence of increasing RVE. If RV "strain" pattern is present, diagnostic study is mandatory as an emergency procedure. Operative mortality is less than one percent, and surgical correction relieves the obstruction.

Aortic Stenosis

Congenital aortic stenosis results in a thrill and ejection systolic murmur in the second right intercostal space. The obstruction may be valvar, subvalvar or supravalvar. Severe obstruction may cause angina, syncope, electrocardiographic evidence of left ventricular strain, and even sudden death. Unfortunately, severe pressure loads on the left ventricle may occur without evidence of symptoms, abnormal ECG or chest film. Indications for diagnostic study are urgent if the clinical diagnosis is clear, and angina, syncope, cardiomegaly or a left ventricular "strain" pattern are present. In the asymptomatic child with a typical murmur and thrill, elective catheterization is indicated after five years of age.

Severity is judged as mild when the peak systolic gradient from left ventricle to aorta is less than 65 mm of mercury, severe when the peak systolic gradient is greater than 80 mm or LV "strain" pattern is present. A moderate category includes those with gradients between 65 and 80 mm of mercury and without an ECG "strain" pattern.

The operation is essentially palliative. Residual aortic stenosis is not uncommon, and late calcific aortic stenosis is common. Aortic insufficiency is not a rare complication, but if mild may be tolerated well through childhood and adolescence. Ultimate valve replacement may be necessary.

Coarctation of the Aorta

Coarctation of the aorta is diagnosed on clinical grounds, by the presence of a differential in blood pressure between the upper and lower extremities. Cardiac catheterization is not indicated unless there is a complicating lesion. Indication for early operation is the presence of severe hypertension, or ECG evidence of left ventricular enlargement. Otherwise, correction should be performed electively between six and twelve years of age. The result is curative.

Tetralogy of Fallot

Tetralogy of Fallot is characterized by a large ventricular septal defect and varying degrees of right ventricular outflow obstruction resulting in RV pressures at or near systemic levels and cyanosis due to right-to-left shunting through the septal defect. The outstanding symptom is decreased exercise tolerance. Sudden decreases in arterial oxygen saturation result in hypoxic spells (increased cyanosis, irritability, and air hunger) and may be lethal. Cardiac catheterization to delineate the anatomy is indicated in all patients in whom operation is contemplated. Two types of surgical approach are available: palliative shunting procedures, or intracardiac repair with closure of the septal defect and relief of outflow tract obstruction.

If the patient is severely symptomatic before the age of five years, a shunting procedure is indicated. Intracardiac repair is indicated in the severely symptomatic child over five years of age, and electively after eight years of age. Palliative shunting procedures in children over five years of age have a low mortality rate and those with good results usually remain satisfactory for five to ten years. Intracardiac repair has been associated with a diminishing mortality rate that is now 7 to 10 percent in many large centers. The long-term results are most promising. Five and ten-year follow-ups have been excellent in those in whom adequate repair has been achieved.